ELSEVIER

Clinical Mass Spectrometry

Subject index

accelerating voltage, 31, 32, 34, 35, 54, 55
acetic acid, 352
acetic anhydride, 93, 99, 116
acetoacetic acid, 192, 196, 197, 298, 299, 304, 315, 327, 329, 337, 342, 361, 367
acetoacetyl-CoA, 329, 332
acetone, 160, 192, 199, 200, 201
acetyl, 91, 92
acetyl derivative, 93, 115, 136
acetylation, 92, 93
acetylcarnitine, 298
acetyl-CFoA, 327
acetylcholine, 99
acetyl-CoA, 199, 200, 303, 306, 307, 315, 318, 319, 324, 325, 327, 329, 332, 348, 362
N^4-acetylcytidine, 131
N-acetylglutamate synthase, 302, 316
N-acetyl-*N*-pentafluorobenzoyl derivative, 99
N-acetyl-*N*-pentafluorobenzoyl derivatization, 99
N-acetylphenylalanine, 300, 301, 335, 339
N-acetyltyrosine, 301, 337, 339, 341
acid hydrolysis, 134
acidosis, 302
ackee, 373
acquired hepatic disease, 9
activated charcoal, 164
active oxygen, 156
acute encephalopathy, 318
acute hepatitis, 265, 266, 277
acute infantile encephalopathy, 318
acylation, 110
acylcarnitine, 8, 293, 321, 367, 368
acyl-CoA, 369
acyl-CoA dehydrogenase, 370
acyl-CoA dehydrogenase deficiency, 365
acylglycine, 294, 308, 367, 368, 369
acyltransferase, 346
adduct ion, 22, 27, 30
adduct ion formation, 20
adenosine, 138
adenosylcobalamin, 311, 312
adenosylcobalamin synthesis deficiency, 311
adipic acid, 159, 192, 193, 196, 197, 199, 282, 295, 296, 297, 299, 301, 319, 322, 329, 351, 353, 354, 361, 365, 366, 367, 368, 370, 371, 372, 373
adipylcarnitine, 296, 367
ado Cbl, 312
adonitol, 179, 205
adrenal cortex, 371
adrenoleukodystrophy, 371, 372
advanced glycation end-products (AGE), 192, 206, 207
Affi-Gel 601 gel, 131
AG-1X formate resin, 133
AGE, 209, 211, 213, 217
alanine, 295, 316, 348, 349
β-alanine, 359
alanine:glyoxylate aminotransferase, 357
albumin-bound metabolite, 163, 164
alcaptonuria, 295, 341, 342
alcohol, 87, 280
aldehyde, 87, 280
aldehyde oxidase, 228
alditol acetate, 94
aldose reductase, 205
aliphatic amine, 156

alkaline hydrolysis, 112
alkaline phosphatase, 134
alkyl phenylpolyethylene sulfate, 53
alkyldimethylsilyl derivative, 110
alkyldimethylsilyl derivatization, 81
alkyldimethylsilylation, 110
alkylphenylpolyethylene sulfate, 49
alopecia, 360
alumina, 95
Amadori, 213
Amadori rearrangement, 206
amine, 5, 95, 96, 97
amino acid, 8, 96, 114, 115, 117, 118, 344
D-amino acid, 115, 117
L-amino acid, 115
amino acid ion exchange chromatography, 215
amino acid sequence, 121, 122, 124, 125
amino acid sequencing, 119, 124
amino acidemia, 117
amino aciduria, 78, 293
amino group, 98
2-aminoadipic acid, 332
2-aminobenzoic acid, 30, 31, 140
2-aminobutyric acid, 296, 355
γ-aminobutyric acid (GABA), 284, 352
S-(2-aminoethyl)-cystein, 120
S-(2-aminoethyl)cysteine residue, 119
aminoguanidine, 217
β-aminoisobutyric acid, 359
5-aminolevulinic acid, 354
α-aminomethylenecyclopropylpropionic acid, 373
ammonia, 20, 22, 40, 254
ammonium acetate, 25, 57
ammonium formate, 57
ammonium ion-adduct ion, 57
amu, 42
p-(*n*-amyl)benzoic acid, 76, 162, 196
amyloidosis, 157
androstanediol glucuronide, 183, 184
androstenediol, 181
androsterone glucuronide, 184
androsterone sulfate, 183, 184
anemia, 161, 170, 361
1,5-anhydroglucitol (1,5-AG), 92, 174, 175, 177, 178, 179, 180, 181, 191, 201, 202, 203, 204
anion association, 41
anion exchange cartridge (SAX), 86, 177, 202, 208
anion exchange chromatography, 111, 282
anion exchange column, 83, 91
anion exchange extraction, 77
anion exchange resin, 92, 164, 204
anthranilic acid, 282
antibody/GC/MS, 103
APCI, 24, 48, 50, 51, 52, 136, 137, 177, 179, 180, 203
APCI-LC/MS, 24, 56, 59, 85, 94, 136, 203
API, 6
apnea, 328, 346
arabinose, 178
arabitol, 161, 175, 178, 179
arginine, 120, 206, 213
arginosuccinic aciduria, 84
aromatic compound, 156
aromatic L-amino acid decarboxylase, 224, 235, 247
arthritis, 341, 342
AST-120 (Kremezin), 174
ataxia, 343, 352, 360, 361
athetosis, 317
atmospheric pressure chemical ionization (APCI), 6, 24, 59
atmospheric pressure ionization (API), 4, 24
atomic mass unit, 42
axial hypotonia, 358
azelaic acid, 159, 282, 299, 301, 308, 313, 366, 372

base, 133, 135, 136, 137
base peak, 16
t-BDMCS, 106, 117, 136
t-BDMS, 110, 286
t-BDMS derivative, 116
t-BDMS derivatization, 105, 106, 116, 136
t-BDMSI, 110
BE, 7, 32, 65, 67
beclomethasone, 111
benzene, 160

benzoic acid, 304
p-(*n*-amyl)benzoic acid, 304
benzoquinoneacetic acid, 343
benzyl alcohol, 173, 174
bile, 253
bile acid, 111, 112, 114, 253, 255, 256, 259, 263, 264, 267, 268, 270, 274, 279, 371, 372
bile acid intermediate, 371
bile acid metabolism, 253, 254, 256
allo(5α[H])-bile acid, 260
bile alcohol, 253, 254, 261, 275, 277, 278
bile salt, 253, 255
bilirubin, 253, 254, 265
biliverdin, 253
binding defect, 169
biopterin, 223, 233
biotin, 302, 304, 311
biotin-dependent carboxylase, 359
biotinidase deficiency, 360
N,*O*-bis(trimethylsilyl)acetamide (BSA), 80
N,*O*-bis(trimethylsilyl)trifluoroacetamide (BSTFA), 76, 80
bleeding tendency, 170
Bond Elut C_{18}, 107, 111, 114
Bond Elut C_{18} cartridge, 113, 275
borate affinity column, 131
boric acid, 94, 95
boric acid gel column, 96
boroacetylation, 94
borohydride, 215
borosilylation, 94
bovine pancreatic ribonuclease A, 130
bovine serum albumin, 125, 127
brain, 96, 97, 99
branched chain 2-keto acid, 346
branched chain amino acid, 194, 346, 348
branched chain amino acidemia, 346
branched chain amino aciduria, 346
branched chain dicarboxylic acid, 366
branched chain hydroxy acid, 349
branched chain keto acid, 346, 348, 349
branched chain α-keto acid dehydrogenase, 345, 346, 348
branched chain ketoaciduria, 346
bromoethylamine, 120
5-bromouridine, 131
Brushfield spot, 371
BSA, 81, 99, 109
BSTFA, 81, 99, 102, 103, 104, 105, 109, 135
BSTFA/TMCS, 78
1-butanol, 201
butanone, 298, 324, 325
2-butanone, 160, 301, 303, 307
n-butyl chloride extraction, 164
n-butyl chloride, 164
butyl ester, 117
butyl ester group, 118
butyl ester-HFB derivative, 116
butyl ester-PFP, 116
n-butyl ester, 115
n-butylboric acid, 94
butylboronate, 177
butylboronate derivatization, 177
butylboronic acid, 177
tert-butyldimethylchlorosilane (*t*-BDMCS), 81
tert-butyldimethylsilyl (*t*-BDMS), 81
tert-butyldimethylsilylation, 81
tert-butyldimethylsilylimidazole (*t*-BDMSI), 81
2-*tert*-butylphenol, 201
butyric acid, 296, 322
butyrylcarnitine, 295, 296
butyryl-CoA, 199, 200
n-butyryl-CoA dehydrogenase, 373
butyrylglycine, 296, 367, 373

C_{10}–C_{14} monoenedicarboxylic acids, 298
C_{27} bile acid, 261, 263
C_{29} dicarboxylic acid, 261, 262
C_{29} dicarboxylic bile acid, 263
C_6–C_{12} dicarboxylic acids, 298
CA, 67
CAD mass spectrum, 66
calcium oxalate, 163, 356, 357
calcium oxalate nephrocalcionosis, 356
calcium oxalate nephrolithiasis, 356
calmodulin, 123
candidiasis, 360
capillary column, 5, 50, 83, 91, 110, 114, 115, 117, 171, 208, 232, 237, 243

capillary column GC/MS, 51, 54, 136, 158, 161, 205
capillary GC column, 50
capillary zone electrophoresis with ESI-MS, 4
capillary zone electrophoresis/mass spectrometry (CZE/MS), 62
carbamoyl phosphate, 363
carbamoyl phosphate synthase I, 302
N-carbamoylamino acid, 364
N-carbamoylaspartic acid, 364
N-carbamoyl-β-alanine, 364
carbohydrate, 139
carbohydrate-deficient glycoprotein syndrome, 128, 129
carbon tetrachloride, 160
carbonic anhydrolase II, 130
3-carboxy-4-methyl-5-ethyl-2-furanpropionic acid, 159, 166, 168
3-carboxy-4-methyl-5-pentyl 2-furanpropionic acid, 159, 166, 168, 169
3-carboxy-4-methyl-5-propyl 2-furanpropionic acid, 159, 164, 166, 167, 168, 169
3-carboxy-5-propyl-2-furanpropionic acid, 159, 166, 168
S-carboxyamidomethylcysteine residue, 119
S-carboxymethylation, 119
S-carboxymethylcysteine residue, 119
1-carboxy-salsolinol, 244
cardiomyopathy, 369, 370, 371
carnitine, 368, 369, 370, 371, 372
L-carnitine, 293, 302, 309, 311, 318, 321, 322, 325, 328, 369
carnitine acyltransferase, 309, 325
L-carnitine deficiency, 294
carnitine palmitoyltransferase (CPT) I deficiency, 368, 370
carnitine palmitoyltransferase (CPT) II deficiency, 370
carnitine transport defect, 368
carnitine/acylcarnitine translocase deficiency, 368, 371
carrier, 55
catalase, 91
cataract, 371
catechol, 160, 170, 171, 172, 173, 174
catecholamine, 95, 96, 98, 99, 223, 224, 244, 246
cation exchange cartridge, 202, 208
cation exchange column, 83, 91, 115
cation exchange resin, 92, 204
cblA, 311
cblB, 311
cblC, 310, 311, 312
cblD, 310, 311, 312
cblF, 310, 311
Cbl^{I}, 312
Cbl^{II}, 312
Cbl^{III}, 312
cerebral atrophy, 350
cerebral dysfunction, 161
cerebral palsy, 318
cerebro-hepato-renal syndrome, 371
cerebrotendinous xanthomatosis (CTX), 260
cesium iodide, 47, 48
CF-FAB, 60, 107
CF-FAB-LC/MS, 107
charcoal adsorption, 133
charcoal-celite, 133
charge exchange, 20, 22, 40, 41
chemical cleavage, 119
chemical ionization (CI), 4, 20
chenodeoxycholic acid, 255, 256, 257, 258, 259, 260, 261, 262, 265, 269, 271, 272
allo-chenodeoxycholic acid, 259, 260
chiral, 239
chiral polysiloxane, 115, 117
chiral stationery phase, 115, 117
chloroform, 100, 160
4-chlororesorcinol, 173
cholanoic acid, 269
5β-cholanoic acid, 256
cholest-4-ene-7α,12α-diol-3-one, 278
cholest-4-ene-7α-ol-3-one, 278
cholest-5-ene-3β,22-diol, 278
cholest-5-ene-3β,24-diol, 278
cholest-5-ene-3β,26-diol, 278
cholest-5-ene-3β,7α-diol, 278
5β-cholestane-3α,7α,12α, 24,26-pentol, 277
5β-cholestane-3α,7α,12α,23,25-pentol glucuronide, 278
5β-cholestane-3α,7α,12α,23,25-pentol, 260, 278

5β-cholestane-3α,7α,12α,24,25,26-hexol, 277
5β-cholestane-3α,7α,12α,24,25-pentol, 277
5β-cholestane-3α,7α,12α,24,26-pentol, 277
5β-cholestane-3α,7α,12α,25,26-pentol, 277
5β-cholestane-3α,7α,12α,25-tetrol glucuronide, 280
5β-cholestane-3α,7α,12α,25-tetrol, 260, 278
5β-cholestane-3α,7α,12α-triol, 260, 261
5β-cholestane-3α,7α-diol, 261
5β-cholestane-3α,7α,12α,23,25-pentol, 278
5β-cholestane-3α,7α,12α,23-tetrol, 278
5β-cholestane-3α,7α,12α,24,25,26-hexol, 278
5β-cholestane-3α,7α,12α,24,25-pentol, 278
5β-cholestane-3α,7α,12α,24,26-pentol, 278
5β-cholestane-3α,7α,12α,24-tetrol, 278
5β-cholestane-3α,7α,12α,25,26-pentol (5β-bufol), 278
cholestane-3α,7α,12α,25-tetrol, 278
5β-cholestane-3α,7α,12α,25-tetrol, 278
5β-cholestane-3α,7α,12α,26,27-pentol, 278
5β-cholestane-3α,7α,12α-triol, 278
5β-cholestane-3α,7α-diol, 278
5α-cholestane-3β,7α-diol, 278
cholestanol, 260, 261
cholestasis, 265, 266, 277
cholestatic, 268
cholestatic liver disease, 114, 279
cholesterol, 199, 255, 260, 261, 302, 303, 311, 361, 362
cholesterol 7α-hydroxylase, 256, 261
cholic acid, 255, 256, 258, 261, 262, 263, 264, 265, 271, 272
allo-cholic acid, 259, 260
cholylglycine hydrolase, 112
chorea, 318
chronic active hepatitis, 275
chronic glomerulonephritis, 155
chronic hepatitis, 266, 271
chronic liver disease, 264
chronic pyelonephritis, 155
chronic skeletal muscle weakness, 369
chymotrypsin, 119, 120
CI, 20, 21, 22, 23, 40, 45, 57, 58, 106, 117, 176, 207, 227
CID, 40, 55, 63, 65, 68, 119, 122, 129, 138
CID mass spectrum, 66
CID-MIKE, 210, 271
CID-MIKES, 314
CIMS, 227, 228, 280
cirrhosis, 277
cis-4-decenoic acid, 369
CI-SIM, 106, 208, 209
citramalic acid, 159, 162
citrate synthase, 305
citric acid, 199, 295, 304, 306, 351
citrulline, 363
cleavage of proteins to peptides, 119
clinical medicine, 7
Clostridium perfringens, 112
clostripain, 119, 120
cluster ion, 27, 64
CNBr, 124
cobalamin reductase deficiency (cblA), 311
cobalamin (cbl), 311, 312
collision, 7
collision cell, 65, 66, 68
collision chamber, 35
collision energy, 66
collisional activation (CA), 65, 66
collisionally-activated dissociation (CAD), 66
collision-induced dissociation (CID), 38, 66
collision-induced dissociation mass analyzed ion kinetic energy (CID-MIKE), 210
coma, 192, 254, 280, 281, 314, 324, 326, 363, 368, 369, 372
conalbumin, 126
congenital biliary atresia, 265
congenital lactic acidosis, 365
conjugated bile acid, 112, 114
conjugated organic acid, 160
conjugated steroid, 184
constant B/E linked scan spectrum, 310
constant B/E ratio, 310
constant B/E ratio-linked scanning, 309
constant neutral loss scan, 65, 67, 68, 117, 118, 263
constant neutral loss spectrum, 68
continuous ambulatory peritoneal dialysis, 133, 158

continuous flow FAB LC/MS (CF-FAB-LC/MS), 6
continuous-flow FAB (CF-FAB), 60
continuous-flow FAB (CF-FAB) LC/MS, 56
convulsion, 334, 346, 348, 358, 363
corneal clouding, 371
corneal erosion, 341
Corona discharge, 24, 59
CPT, 370
CPT II deficiency, 368
craniofacial dysmorphia, 371
creatine kinase, 361
creatinine, 156, 158
p-cresol, 160, 170, 172, 173, 174, 282
crotonase, 327
crotonoyl-CoA, 332
crotonyl-CoA, 319
CsI, 50
CTX, 261, 278, 280
cyanogen bromide (CNBr), 120
cyanosis, 328
cyclohexanone, 160, 201
cyclopropane-ring carboxylic acid, 282
cysteine, 120, 344
cysteinylglycine, 343, 344
cystic fibrosis, 271, 273
cytidine, 135, 138
cytochrome P450, 228, 238
cytosine, 359
CZE, 16, 62, 157
CZE/CF-FAB-MS, 63
CZE/ESI-MS/MS, 63
CZE/MS, 56

daughter ion, 63
DC voltage, 33, 34, 37
DCI-MS, 158
DEAE cellulose, 133
DEAE column, 133
DEAE-Sephadex, 77
DEAE-Sephadex A-25, 108
DEAE-Sephadex A-25 anion exchange column, 109
DEAP-LH-20, 111
death, 372
debrisoquine, 238
decanoylcarnitine, 296
decanoylglycine, 300, 367, 369
decarboxylase, 346
decenedioic acid, 295, 370, 373
2-decenedioic acid, 366
3-decenedioic acid, 366
4-decenedioic acid, 366
5-decenedioic acid, 366
decenoylcarnitine, 367, 368
deconvoluted spectrum, 30
5-decynedioic acid, 159
5,6-decynedioic acid, 282
dehydration, 192, 310
dehydroascorbic acid, 213
dehydroepiandrosterone glucuronide, 183
dehydroepiandrosterone sulfate, 182, 184
11-dehydro-TXB_2, 106
delayed motor development, 358
3-deoxy-2-(hydroxymethyl)tetronic acid, 162
3-deoxy-2C(hydroxymethyl)tetronic acid, 159
deoxyadenosine, 138
deoxyalditol, 177
6-deoxyallitol, 175, 176
5-deoxyarabitol, 175, 176
deoxycholic acid, 255, 256, 260, 269, 272
deoxycytidine, 138
4-deoxyerythritol, 175, 176
4-deoxyerythronic acid, 159, 162, 199
2-deoxyerythropentonic acid, 159
3-deoxyfructose, 209
N^{ε}-(1-deoxyfructosyl), 217
6-deoxygalactitol, 175, 176
N^{ε}-(1-deoxyglucitolyl)lysine, 215, 217
1-deoxyglucose, 92, 177, 201
3-deoxyglucosone, 206, 207, 208, 209, 213, 217
deoxyguanosine, 138
2-deoxyguanosine, 136
6-deoxygulitol, 175, 176
3-deoxyhexitol, 209
N^{ε}-(1-deoxyhexitolyl)lysine, 217
6-deoxymannitol, 175, 176
deoxynucleoside, 138, 139
2-deoxyribitol, 175, 176
deoxyribonucleoside, 136

deoxyribonucleotide, 139, 140
2-deoxytetronic acid, 162
3-deoxytetronic acid, 159, 162
2-deoxytetrono-1,4-lactone, 162
4-deoxythreitol, 175, 176
4-deoxythreonic acid, 159, 162, 199
5-deoxyxylitol, 175, 176
deprenyl, 226
deproteinization, 79, 87, 164
N-desmethyldiazepam, 285, 286
desolvator temperature, 179
desorption chemical ionization (DCI), 4, 23, 24
detector, 15
developmental delay, 350, 360
developmental retardation, 302, 310, 311
DHCA, 371
DHPG, 246
1,1-di(4-chlorophenyl)-2,2-dichloroethene, 160
3,5-di(trifluoromethyl)benzoyl chloride (DTFMBCl), 99
3,5-di(trifluoromethyl)benzoyl-isopropyl-dimethylsilyl (DTFMB-IPDMS), 99
3,5-di(trifluoromethyl)benzoyl-TMS (DTFMB-TMS), 99
diabetes, 204
diabetes mellitus, 6, 9, 91, 175, 191
diabetic ketoacidosis, 191, 192, 193, 194, 197, 199, 200
diabetic ketoacidotic urine, 196, 199
diabetic nephropathy, 155, 191, 209
diabetic neuropathy, 191, 205
diabetic retinopathy, 191
diagnosis of disorders of organic acid metabolism, 8
dialysate, 158
diarrhea, 361, 372
diazepam, 285, 286
diazomethane, 82, 105, 113, 169
dicarboxylic acid, 161, 192, 199, 262, 367, 369, 373
dicarboxylic aciduria, 365, 367, 368
2,3-dideoxypentonic acid, 196
2,4-dienoyl-CoA reductase deficiency, 368, 371
diethyl ether, 76, 87, 89, 100, 107
diethylaminoethyl bromide (DEAEB), 107
diethylaminoethyl chloride (DEAEC), 107
diethylaminoethyl derivatization, 107
diethylaminohydroxypropyl-LH-20 (DEAP-LH-20), 109
digit-printing method, 123
1,2-dihydroisoquinoline, 237, 239
dihydrolipoyl dehydrogenase (E_3) deficiency, 295, 347
dihydrolipoyl dehydrogenase deficiency, 347
dihydropyrimidine dehydrogenase, 358, 359
dihydropyrimidine dehydrogenase activity, 359
dihydropyrimidine dehydrogenase deficiency, 295, 358, 359
5,6-dihydropyrimidine, 135
5,6-dihydrothymine, 359
5,6-dihydrouracil, 358, 359
dihydrouridine, 135
6,7-dihydroxy-1,2,3,4-tetrahydroisoquinoline, 244
3α,7α-dihydroxy-12-keto-5β-cholanoic acid, 270
2,3-dihydroxy-2-methylbutyric acid, 304
7α,12α-dihydroxy-3-keto-4-cholenoic acid, 258, 259, 260
7α,12α-dihydroxy-4-cholesten-3-one, 259
3α,7α-dihydroxy-5β-cholestanoic acid, 261, 371
3β,12α-dihydroxy-5-cholenoic acid, 266
3β,7α-dihydroxy-5-cholenoic acid, 256, 266
3β,7β-dihydroxy-5-cholenoic acid, 266
3α,12α-dihydroxy-7-keto-5β-cholanoic acid, 270
dihydroxyacetone phosphate, 351
dihydroxyacetone phosphate acyltransferase, 262
2,5-dihydroxybenzoic acid (DHBA), 30, 31
2,4-dihydroxybutyrate lactone, 297
3,4-dihydroxybutyrate lactone, 297
2,4-dihydroxybutyric acid, 297, 353, 354
3,4-dihydroxybutyric acid, 297, 352, 353, 354
dihydroxycholanoate, 273
dihydroxycholanoate sulfate, 273

dihydroxycoprostanic acid (DHCA), 261, 262
4,5-dihydroxyhexanoate lactone, 297
4,5-dihydroxyhexanoic acid, 297, 353
3,4-dihydroxymandelic acid, 96
3,4-dihydroxyphenylacetaldehyde, 245
3,4-dihydroxyphenylacetic acid, 96
L-3,4-dihydroxyphenylalanine, 223
3,4-dihydroxyphenylethanol (DHPE), 246
3,4-dihydroxyphenylethyleneglycol (DHPG), 246
3,4-dihydroxyphenylpyruvic acid, 245
L-threo-3,4-dihydroxyphenylserine, 223
diisopropylethylamine, 104
4,6-diketoheptanoic acid, 339
3,5-diketooctanedioic acid, 339
dilichol, 362
3,5-dimethoxy-4-hydroxy-*trans*-cinnamic acid, 129
dimethyl sulfoxide (DMSO), 135
1,2-dimethyl-6,7-dihydroxy-1,2,3,4-tetrahydroisoquinoline, 241
1,2-dimethyl-6,7-dihydroxy-isoquinolinium ion ($DDIQ^+$), 244
2,4-dimethyladipic acid, 159
dimethylamine, 156
dimethylethylpyrazine, 201
dimethylethylsilyl (DMES), 81
dimethylethylsilylimidazole (DMESI), 82
N,*N*-dimethylformamide (DMF, 116
N^2,N^2-dimethylguanine, 131
N^2,N^2-dimethylguanosine, 131
dimethyl-i-propylsilylimidazole (DMiPSI), 82
dimethylisopropylsilyl (DMiPS), 81
dimethyl-*n*-propylsilyl (DMnPS), 81
dimethyl-*n*-propylsilylimidazole (DMnPSI), 82
2,3-dimethylpyrazine, 201
dimethyltrisulfide, 201
2,4-dinitrophenyl (DNP) amino acid methyl ester, 117
26,27-dinor-5β-cholestane-3α,7α,12α,24,25-pentol, 278
2,3-dinor-6-ketoPG$_{1\alpha}$, 103
2,3-dinor-6-keto-PGF$_{1\alpha}$, 102
2,3-dinor-TX, 101
2,3-dinor-TXB$_2$, 101, 102, 103, 105, 106
diol, 101
cis-diol, 95, 96, 131, 177
2,6-diphenyl-*p*-phenyleneoxide, 88
dipropylketone, 160
direct coupling, 51
direct injection, 86, 87, 88, 91
direct liquid introduction (DLI), 136
discharge electrode, 57
discharge-assisted TSI, 57
disialotransferrin, 129
disordered coagulation, 161
disorders of organic acid metabolism, 6
dissociative electron capture, 41
dithiothreitol (DTT), 119
DLI-LC/MS, 137
DMES, 110, 113, 266, 267, 268, 269, 270
DMESI, 110
DMF, 116
DMiPS, 105, 110
DMiPS derivatization, 106
DMiPSI, 106, 110
DMnPS, 110
DMnPSI, 110
DNA, 134, 136, 140
DNase I, 134
DNP amino acid, 117
docosahexaenoic acid, 106
dodecanedioic acid, 366, 373
dodecanedioylcarnitine, 296
dodecanoylcarnitine, 367
dodecenedioic acid, 373
3-dodecenedioic acid, 366
5-dodecenedioic acid, 366
DOPA, 224
L-DOPA, 96, 223, 224, 225, 233, 243, 244, 245, 246
dopamine, 223, 224, 226, 228, 230, 233, 235, 243, 244, 245, 246
dopamine β-hydroxylation, 224
dopamine β-hydroxylase, 223
L-DOPS, 96, 223, 246, 247
L-threo-DOPS, 224
dose-dependent manner, 181
double-focusing magnetic sector electric sector mass analyzer, 4

double-focusing mass spectrometer, 32, 33, 65
Dowex 50W-X4, 115
Dowex 50-X8, 115
drift voltage, 59
drug metabolism, 5
DTFMBCl, 99
dysmorphic feature, 261, 361
dystonia, 317

E_1, 346
$E_{1\beta}$ deficiency, 346
E_2, 346
E_2 deficiency, 346
E_3, 346, 348
E_3 deficiency, 346, 348, 349
EB, 32, 33, 65, 67
eczema, 334
Edman degradation, 124
EI, 19, 40, 44, 58, 79, 80, 92, 93, 94, 98, 106, 168, 172, 176, 195, 215, 217
eicosanoid, 106, 107
EI-MS, 105, 121
EIMS, 215, 217
electrode, 19
electron capture, 40, 41
electron capture detector (ECD), 5, 98
electron filament, 57
electron ionization (EI), 3, 16, 18
electron transfer flavoprotein (ETF) deficiency, 368
electron trap, 19
electrophilic addition, 22
electrophilic compound, 41, 42
electrophilic group, 40, 41
electrospray ionization (ESI), 4, 28
electrospray ionization (ESI) LC/MS, 61
electrostatic field, 67
electrostatic field strength, 33
electrostatic sector, 32, 33, 65, 210
element, 43
elemental composition, 33, 42, 44, 45
emitter, 23, 24
enantiomer, 117
enantiomer-labelling, 115
encephalomyopathy, 350
encephalopathy, 317, 322, 355, 373
endcap electrode, 37
endogenous digitalis-like factor, 182
end-stage renal disorder, 155
energy spectrum, 65
enoylhydratase, 327
enterohepatic circulation, 256
enzymatic cleavage, 119
enzymatic digest, 125
enzymatic digestion, 119
enzymatic hydrolysis, 112
enzyme hydrolysis, 134
EO, 83
EO-TMS, 83
epiandrosterone glucuronide, 184
epiandrosterone sulfate, 183, 184
epinephrine, 223, 224, 245, 246
epinine, 96
epinine diisobutyric ester, 99
erythritol, 161, 175, 177, 178, 179, 180
erythro-4,5-dihydroxyhexanoic acid, 354
erythroid colony formation (CFU-E), 169
erythronic acid, 159
erythropoiesis, 156
ESI, 8, 30, 49, 52, 53, 61, 64, 119, 125, 128
ESI mass spectrum, 126, 127
ESI-LC/MS, 6, 8, 56, 62, 121, 125
ESI-LC/MS/MS, 8, 121
ESI-MS, 119, 125, 127, 129, 139, 156, 157
ESI-MS/MS, 139
estrogen, 108
ETF, 322, 324, 370
ETF:QO, 322, 370
ETF:ubiquinone oxidoreductase (ETF:QO) deficiency, 368
1-ethanol, 201
ethoximation, 83
ethoxime, 83
N-ethoxycarbonyl derivative, 98
ethyl acetate, 76, 100, 107, 116
3-ethyl-3-hydroxyglutaric acid, 301, 303, 307
3-ethyl-3-hydroxyglutaryl-CoA, 307
2-ethyl-3-ketohexanoic acid, 200
2-ethylacrylyl-CoA, 194
ethylchloroformic acid, 98
ethylene glycol, 314
ethyleneimine, 119, 120

2-ethylhydracrylic acid, 162
O-ethylhydroxylamine hydrochloride, 83
ethylmalonic acid, 282, 295, 296, 322, 323, 366, 370, 373
ethylmalonic acid-adipic aciduria, 295, 370
ethyloxime, 83
p-ethylphenol, 173
ethyl-triphenylphosphonium derivatization, 122
even-numbered dicarboxylic acid, 372
exact mass, 42, 43, 44, 45, 50, 52, 53
extrahepatic cholestasis, 258
extrahepatic obstructive jaundice, 264

FAB, 6, 8, 23, 24, 25, 26, 46, 47, 48, 50, 64, 93, 136, 183, 216, 257, 310
FAB-CID-MS/MS, 213, 214
FAB-LC/MS, 121
FAB-LC/MS/MS, 121
FAB-MS, 27, 41, 46, 64, 99, 114, 117, 119, 120, 121, 123, 125, 137, 138, 139, 156, 182, 183, 213, 217, 258, 259, 261, 263, 271, 273, 278, 280, 293, 309, 321, 325
FAB-MS/MS, 75, 117, 121, 122, 261, 262, 273, 294, 310, 364
FAD, 318, 319, 346
familial amyloid polyneuropathy, 246
familial amyloidotic polyneuropathy, 8
familial giant-cell hepatitis, 258, 259
Fanconi syndrome, 337
farnesylpyrphosphoric acid, 362
fast atom bombardment (FAB), 4, 25
FD, 23
ferrulic acid, 140
FFI, 211
FFR, 68
field desorption (FD), 4, 23
field free region (FFR), 67
field ionization (FI), 23
filament, 18
flame ionization detector (FID), 5
flavin-containing monooxygenase, 228
flavoprotein, 322
1-fluoro-(2,4)-dinitrobenzene (FDNB), 117
fluoroacyl anhydride, 97, 98
N-fluoroacylimidazole, 97
forward geometry, 33
forward geometry double-focusing mass spectrometer, 67
forward geometry instrument, 68
Fourier transform (FT), 38
Fourier transform mass spectrometry (FTMS), 38
Fourier transform-ion cyclotron resonance mass spectrometry (FT-ICRMS), 38
four-sector mass spectrometer, 121
four-sector tandem mass spectrometer, 68
four-sector tandem MS, 65, 68
fractures, 351
fragment ion, 15, 16, 17, 18, 25, 27, 42, 44
fragmentation, 17, 27, 58
free organic acid, 158
frit target, 60
frit-FAB, 107
frit-FAB-LC/MS, 6, 56, 60, 61, 99, 125, 127, 131, 133
fructosamine, 206
fructose, 175, 178, 205, 213, 358
fructose 1,6-bisphosphatase deficiency, 349
FT, 125
FTMS, 40, 63, 65, 68
fumarase, 349, 350
fumarase deficiency, 350, 351
fumaric acid, 162, 295, 304, 337, 342, 349, 350
fumaric aciduria, 349, 350
fumaric aciduria (fumarase deficiency), 295
fumarylacetoacetase, 337, 338
fumarylacetoacetic acid, 337, 338, 342
furan, 87
furancarboxylic acid, 169
furosine, 211
furoyl, 210, 211
2-(2-furoyl)-4(5)-(2-furanyl)-1H-imidazole (FFI), 209
furoylglycine, 160

β-galactose, 175, 178
gallstone, 114
gas chromatography (GC), 5
gas chromatography/mass spectrometry (GC/MS), 50
gas-phase stripping, 89
GC, 7, 8, 86, 91, 93

GC/MS, 6, 7, 8, 54, 55, 75, 76, 83, 86, 91, 92, 93, 94, 96, 97, 98, 99, 102, 103, 104, 105, 106, 108, 109, 110, 112, 113, 114, 115, 121, 134, 135, 136, 156, 157, 158, 164, 169, 170, 176, 177, 182, 191, 192, 194, 195, 199, 200, 201, 207, 208, 209, 224, 225, 227, 228, 230, 231, 236, 242, 243, 244, 245, 246, 247, 259, 261, 264, 265, 266, 273, 280, 281, 282, 284, 285, 293, 294, 312
GC/NICIMS, 102, 239, 266, 270, 286
GC/NICIMS/MS, 106
GC/SIM, 98
gel permeation chromatography, 157, 158
Gelpack GL-C64Z column, 179
geranylpyrphosphoric acid, 362
glaucoma, 371
globin, 119
glomeruli, 157
gluconate, 91
glucopyranurono-(6-1)-lactone, 304
glucose, 91, 92, 179, 191, 205, 206, 209, 210, 213, 216, 217
α-glucose, 175, 178
β-glucose, 175, 178
D-glucose, 211
glucose 6-phosphatase deficiency, 349
glucose oxidase, 91
glucuronate-xylulose-pentose pathway, 181
glucuronic acid, 316
D-glucuronic acid, 181
β-glucuronidase, 113
β-glucuronidase/sulfatase, 108, 109
glucuronidated bile acid, 265
glucuronide, 99, 107, 157, 170, 174, 181, 260, 369
glucuronide-conjugated bile acid, 111, 113
glutaconic acid, 296, 319, 320
glutaconyl-CoA, 318, 319
glutaconyl-CoA decarboxylase, 319
glutamic acid, 120, 295, 316, 344, 348, 349, 352
glutamic-oxaloacetic transaminase (GOT), 253
glutamic-pyruvic transaminase (GPT), 253
glutamine, 295, 348, 349
L-2-keto acid aminotransferase, 284
γ-glutamylamino acid, 335, 343, 344
γ-glutamyl cycle, 284, 335, 343
γ-glutamylcyclotransferase, 284, 344
γ-glutamylcysteine, 344
γ-glutamylcysteine synthetase, 344
L-γ-glutamyl-L-cysteinylglycine, 343
γ-glutamylphenylalanine, 300, 335
γ-glutamyltransferase, 335, 343, 344
γ-glutamyl transpeptidase (γ-GTP), 264
glutaric acid, 159, 282, 296, 297, 298, 301, 304, 308, 318, 319, 322, 329, 332, 353, 366, 373
glutaric acidemia type 1, 317
glutaric acidemia type 2, 322
glutaric aciduria type 1, 294, 296, 317, 318, 319, 321
glutaric aciduria type 2, 296, 324, 365, 370
glutarylcarnitine, 296, 321, 322
glutaryl-CoA, 308, 318, 319, 332
glutaryl-CoA dehydrogenase, 308, 317, 318, 319, 322, 370
glutaryl-CoA dehydrogenase activity, 373
glutathione, 343, 344
glutathione deficiency, 343
glutathione *S*-transferase, 169
glutathione synthetase, 344
glutathione synthetase deficiency, 343
glycated albumin, 206, 210
glycation, 206, 215, 216
glycation of protein, 192
D-glycerate dehydrogenase, 357
D-glycerate kinase deficiency, 358
glyceric acid, 159, 162
D-glyceric acid, 296, 358
L-glyceric acid, 163, 300, 356, 357
D-glyceric aciduria, 296, 357, 358
L-glyceric aciduria, 300, 356
glycerol, 26, 27, 56, 60, 61, 162, 174, 175, 296, 310, 351
glycerol kinase activity, 351
glycerol-3-phosphate, 351
glycerolipid, 351
glyceroluria, 296, 351
glycine, 255, 256, 259, 315, 327, 344, 369
glycine cleavage enzyme, 302
glycine conjugate, 112

glycine *N*-acylase, 315, 316, 324
glycine-conjugated bile acid, 111
glycoaldehyde, 217
glycochenodeoxycholate, 255
glycochenodeoxycholic acid, 275, 279
glycocholate, 255
glycocholic acid, 275, 279
glycodihydroxy, 273
glycodihydroxycholanoate, 114
glycodihydroxycholanoate sulfate, 114
glycodihydroxysulfate, 273
glycogen storage disease type 1, 349, 365, 373
glycogen storage disease type 3, 365, 373
glycolic acid, 162, 297, 300, 352, 354, 355, 357
glycolic aciduria, 300, 356
glycomonohydroxy, 273
glycomonohydroxycholanoate, 114
glycosylated amino acid, 121
glycosylation, 130
glycotetrahydroxy, 273
glycotetrahydroxycholanoate, 114
glycotrihydroxy, 273
glycotrihydroxycholanoate, 114
glyoxylic acid, 300, 356, 357
GOT, 264
GPT, 264
granddaughter ion, 68
growth retardation, 358
guanidine, 156
guanidine compound, 156
guanidinosuccinic acid, 156
guanosine, 135, 138
Guthrie card, 117

Hb A_2 Honai, 124
headspace sampling, 88
Helix promatia, 109
heme, 354
heme A, 362
hemodialysate, 161
hemodialysis, 94, 133, 156, 158, 161, 163, 169, 170, 173, 180
hemofiltration, 182
hemoglobin, 115, 206
hemoglobin A_{1C}, 215
hemoglobin variant, 8, 123
hemolytic anemia, 343
hemolytic effect, 156
hepatic disease, 253
hepatic dysfunction, 369
hepatic encephalopathy, 254, 280, 281, 284, 285, 286
hepatobiliary disease, 263, 264
hepatomegaly, 261, 318, 322, 328, 369, 371
hepatosplenomegaly, 361
n-heptadecanoic acid, 77
heptafluorobutyl (HFB), 97
heptafluorobutyric anhydride (HFBA), 97
heptafluorobutyrylimidazole (HFBI), 97
2-heptanone, 200, 201
4-heptanone, 160, 200, 201
3-heptene-2-one, 201
heptenedioic acid, 301, 366, 371
hereditary tyrosinemia, 337
12-HETE, 106
15-HETE, 106
5-HETE, 106
hexachlorobenzene, 160
hexadecanedioic acid, 366, 372, 373
hexadecenedioic acid, 366, 373
hexafluoroisopropanol, 113, 116
hexafluoroisopropyl (HFIP), 113
hexamethyldichlorosilane, 113
hexamethyldisilazane (HMDS), 80
hexanoylcarnitine, 295, 296, 367, 368
hexanoyl-CoA, 199
hexanoylglycine, 295, 296, 300, 324, 367, 368, 370, 373
hexenedioic acid, 366
4-hexenoic acid, 373
HFB, 98, 115, 230, 231, 232, 233, 234, 235, 238, 240, 241
HFB derivatization, 110
HFBA, 97, 116
HFBI, 98, 110
HFIP derivatization, 114
HFIP ester, 115
HFIP-PFP, 116
HFIP-TFA, 264, 265
HFIP-TFA derivatization, 113
high energy CID, 66
high resolution, 66, 138

high resolution CIMS, 45
high resolution FAB-MS, 48, 213
high resolution magnetic mass spectrometer, 125
high resolution mass spectrometry (HRMS), 42
high resolution MS, 32, 33, 42, 44, 48, 124, 184
high resolution selected ion monitoring (HR-SIM), 55
hippuric acid, 156, 158, 160, 164, 304
HMDS, 109
HMDS/TMCS, 110
HMG-CoA reductase, 260, 362
HO, 82
holocarboxylase synthetase deficiency, 360
homocatechol, 160, 173, 174
homocitric acid, 306
homocystine, 299
homocystinuria, 312
homogentisic acid, 295, 337, 342, 343
homogentisic acid oxidase, 282, 341, 342
homogentisic acid polyphenol oxidase, 342
homogentisic aciduria, 341
homoserine lactone residue, 120
homovanillic acid, 160, 282
HPLC, 16, 104, 114, 131, 136, 157, 158, 164, 169, 182, 213, 215, 217, 226, 227, 228
HRGC/NICIMS, 104, 105
HRSIM, 106, 110, 114
hybrid geometry instrument, 35
hybrid type, 65
hybrid type MS/MS, 66
hydride abstraction, 20
hydrogen flame ionization detector, 98
2-hydroisovaleric acid, 349
hydrophobic resin, 164
hydroquinone, 160, 170, 171, 172, 173, 174
hydroxime, 82
ψ-hydroxy acid, 369
hydroxy fatty acid, 106
3β-hydroxy-Δ^5-C_{27}-steroid dehydrogenase deficiency, 256
3-hydroxy-1,10-decandioic acid, 195
3-hydroxy-1,12-dodecanedioic acid, 195
3α-hydroxy-12-keto-5β-cholanoic, 270
4-hydroxy-2-butenoic acid, 352
3-hydroxy-2-ethylpropionic acid, 193, 194
3-hydroxy-2-ethylpropionyl-CoA, 194
2-hydroxy-2-methylbutyric acid, 162
3-hydroxy-2-methylbutyric acid, 192, 193, 194
3-hydroxy-2-methylbutyryl-CoA, 194
2-hydroxy-2-methyllevulinic acid, 193, 195, 196, 197, 199
2-hydroxy-2-methylsuccinic acid, 300, 303, 304, 306
7α-hydroxy-3-keto-4-cholenoic acid, 258, 259, 260
4-hydroxy-3-ketobutyric acid, 297, 352, 353, 354
4-hydroxy-3-methoxyphenylpropionic acid, 282
3-hydroxy-3-methylglutaconyl-CoA, 315
3-hydroxy-3-methylglutaric acid, 199, 297, 301, 304, 327, 329, 330, 356
3-hydroxy-3-methylglutaric aciduria, 84, 297, 327, 328, 329, 330, 356
3-hydroxy-3-methylglutaryl-CoA (HMG-CoA) reductase, 361
3-hydroxy-3-methylglutaryl-CoA lyase, 328, 329
3-hydroxy-3-methylglutaryl-CoA, 199, 315, 327, 329, 356, 362
2-hydroxy-3-methylvaleric acid, 193, 194, 298, 346, 347
7α-hydroxy-4-cholesten-3-one, 259
3β-hydroxy-5-cholenoic acid, 265, 266
3β-hydroxy-5-ene metabolite, 266
3β-hydroxy-5-en-unsaturated bile acid, 266
3α-hydroxy-7,12-diketo-5β-cholanoic acid, 270
4(or 5)-hydroxy-5(or 4)-ketohexanoic acid 5)-, 353
8-hydroxyadenine, 136
2-hydroxyadipic acid, 298, 332, 333
hydroxyalkoxypropyl Sephadex, 109
4-hydroxyandro-4-ene-3,17-dione, 111
11β-hydroxyandrosterone, 161, 181
3-hydroxyazelaic acid, 366, 369
2-hydroxybenzoate glucuronide, 160
3-hydroxybenzoic acid, 159

4-hydroxybenzoic acid, 156, 159, 161, 162, 170, 282
2-hydroxybutyric acid, 193, 196, 197, 199, 295, 296, 298, 299, 349, 355, 361, 367
3-hydroxybutyric acid, 192, 196, 197, 298, 299, 304, 313, 361, 367, 373
4-hydroxybutyric acid, 297, 352, 353, 354
2-hydroxybutyric aciduria, 296, 355
4-hydroxybutyric aciduria, 297, 352, 353, 354
3-hydroxybutyryl-CoA, 332
ψ-hydroxycarboxylic acid, 367
2-hydroxycarboxylic acid, 194, 367, 372
3-hydroxycarboxylic acid, 198, 367
7α-hydroxycholesterol to 7α-hydroxy-4-cholesten-3-one, 256
26-hydroxycholesterol, 265
3-hydroxydecanedicarboxylic acid, 193, 194, 195
9-hydroxydecanedioic acid, 367
9-hydroxydecanoic acid, 300, 368
3-hydroxydecenedicarboxylic acid, 193, 194
3-hydroxydicarboxylic acid, 366, 369
2-hydroxydicarboxylic acid, 366
3-hydroxydihydrofuran-2(3H)-one, 297, 353, 354
4-hydroxydihydrofuran-2(3H)-one, 297, 353, 354
3-hydroxydodecanedicarboxylic acid, 193, 194, 195
3-hydroxydodecanedioic acid, 366, 369
3-hydroxydodecenedicarboxylic acid, 193, 194
hydroxyeicosatetraenoic acid (HETE), 100
5-(1-hydroxyethyl)dihydrofuran-2(3H)-one, 297, 354
11β-hydroxyetiocholanolone, 161, 181
D-2-hydroxyglutaric acid dehydrogenase, 354
2-hydroxyglutaric acid, 159, 162, 295, 296, 297, 304, 322, 323, 324, 348, 349, 354, 355, 366, 372
3-hydroxyglutaric acid, 296, 319, 320
D-2-hydroxyglutaric acid, 354
2-hydroxyglutaric aciduria, 297, 354
8-hydroxyguanine, 136
3-hydroxyhexadecanedioic acid, 366, 370
3-hydroxyhexanoic acid, 193, 195, 197, 304, 367
5-hydroxyhexanoic acid, 193, 195, 196, 197, 199, 299, 304, 367, 368, 373
2-hydroxyhippuric acid, 157, 160, 164
3-hydroxyhippuric acid, 160, 164
4-hydroxyhippuric acid, 160, 164, 282
2-hydroxyisobutyric acid, 296, 322, 367
3-hydroxyisobutyric acid, 162, 192, 193, 194
3-hydroxyisobutyryl-CoA, 194
3-hydroxyisoheptanoic acid, 297, 315, 317
2-hydroxyisohexanoic acid, 193, 194, 295, 298, 346, 347, 348, 349, 372
2-hydroxyisovaleric acid, 193, 194, 196, 197, 295, 298, 346, 347, 349, 367
3-hydroxyisovaleric acid, 192, 193, 194, 196, 197, 296, 297, 298, 299, 301, 304, 308, 315, 316, 317, 322, 327, 329, 356, 361, 367
4-hydroxyisovaleric acid, 297, 315, 316
3-hydroxyisovaleryl-CoA, 194, 315, 327, 329
4-hydroxyisovaleryl-CoA, 315
hydroxyl group, 98
hydroxylamine hydrochloride, 77, 83
hydroxylysine, 206, 308, 318, 319, 332
4-hydroxymandelic acid, 160, 282
5-hydroxymethyl-1-alkylpyrrole-2-carbaldehyde, 211
5-hydroxymethyluracil, 295, 358, 359
3-hydroxymyristic acid, 77
3-hydroxyoctanedicarboxylic acid, 193, 194
3-hydroxyoctanoic acid, 193, 195, 197, 198, 367
7-hydroxyoctanoic acid, 300, 367, 368
3-hydroxyoctenedicarboxylic acid, 193, 194
4-hydroxyphenolic acid, 281
3-(3-hydroxyphenyl)-3-hydroxypropionic acid, 160, 164
2-hydroxyphenylacetic acid, 85, 86, 159, 300, 301, 334, 335
3-hydroxyphenylacetic acid, 159
4-hydroxyphenylacetic acid, 156, 159, 161, 162, 164, 170, 282, 297, 299, 301, 304, 337, 339, 341, 361, 367
4-hydroxyphenylacetic aciduria, 297

2-(4-hydroxyphenylazo)-benzoic acid (HABA), 30, 31, 129
m-hydroxyphenylethanol (mHPE), 246
p-hydroxyphenylethanol (pHPE), 246
p-hydroxyphenylglycol, 246
4-hydroxyphenyllactic acid, 282, 301, 304, 337, 338, 339, 341
4-hydroxyphenylpropionic acid, 282
4-hydroxyphenylpyruvic acid oxidase, 282
4-hydroxyphenylpyruvic acid, 282, 301, 336, 337, 339, 341, 342
3-hydroxypicolinic acid, 30, 31, 139
3-hydroxypropionic acid, 162, 297, 299, 300, 303, 304, 305, 306, 313, 325, 327, 330, 353, 354, 361
3-hydroxypropionyl-CoA, 353
hydroxypyruvic acid, 357
2-hydroxysebacic acid, 262, 366, 372
3-hydroxysebacic acid, 366, 369, 372
3-hydroxysuberic acid, 366, 369
3-hydroxytetradecadienedicarboxylic acid, 193, 195
3-hydroxytetradecanedioic acid, 366, 370
3-hydroxytetradecenedicarboxylic acid, 193, 195
1β-hydroxy-THCA, 262
25-hydroxy-THCA, 262
26-hydroxy-THCA, 262
6α-hydroxy-THCA, 262
4-hydroxy-TIQ, 238
3-hydroxytridecanedioic acid, 366, 369
1β-hydroxyursodeoxycholic acid, 271
21-hydroxyursodeoxycholic acid, 271, 272
3-hydroxyvalereric acid, 303
3-hydroxyvaleric acid, 193, 195, 196, 197, 199, 299, 300, 304, 307, 313, 361
4-hydroxyvaleric acid, 304
hyocholic acid, 259, 262, 269, 272
hyperammonemia, 302, 310, 322, 324, 328, 363, 368
hyperammonemic coma, 369
hyperglycerolemia, 351
hyperglycinemia, 302, 310, 358
hyperkeratosis, 341
hyperkinesis, 352
hyperpipecolic acidemia, 261
hypertrophic cardiomyopathy, 369
hypoglycemia, 302, 310, 318, 322, 326, 328, 346, 348
hypoglycemic encephalopathy, 370
hypoglycine A, 373
hypoketotic hypoglycemia, 368, 369, 370
hypotonia, 261, 311, 317, 326, 330, 348, 350, 352, 358, 360, 371
hypoxanthine, 133, 158

ibopamine, 99
imino group, 98
immunoaffinity column, 104
immunoaffinity column extraction, 100
immunoaffinity extraction, 76, 103
immunoreactive digitalis-like factor, 184
impairment of nervous system, 170
inborn error of bile acid metabolism, 9
indole, 201
indole amine, 98
indole-3-acetic acid, 164, 336
indoleacetic acid, 282
indolic compound, 156
indoxyl sulfate, 156, 158, 159, 164
infantile Refsum's disease, 261, 262
infrared (IR) spectroscopy, 7
inherited metabolic disorder, 8
inhibition of brain enzymes, 161
inhibition of platelet aggregation, 161
inhibitor of albumin binding of drugs, 169
inositol isomer, 181
chiro-inositol, 161, 175, 176, 177, 178, 181
cis-inositol, 175
epi-inositol, 175
myo-inositol, 161, 174, 175, 176, 177, 178, 179, 180, 181, 201, 205
neo-inositol, 175
scyllo-inositol, 161, 175, 176, 177, 178, 181
insulin-dependent, 204
insulin dependent diabetes mellitus (IDDM), 1191, 202
interface, 50
internal standard, 55, 178
intrahepatic cholestasis, 267
iodoacetamide, 119
iodoacetic acid, 119
ion cyclotron resonance, 38

ion source, 15
ion spray, 29, 62
ion trap (IT), 37, 125
ion trap (IT) mass spectrometer, 4
ion trap mass spectrometry (ITMS), 37
ion-molecule reaction, 20, 24, 25, 29, 38, 57, 59
ion-pair production, 41
isobutane, 20, 21, 22, 40
isobutyl ester, 115
isobutylchloroformic acid, 98
N-isobutyloxycarbonyl derivative, 98
isobutyric acid, 296, 322
isobutyrylcarnitine, 295
isobutyryl-CoA, 194, 346
isobutyrylglycine, 296, 324, 367
isocyanate, 88
isoleucine, 192, 194, 199, 254, 280, 295, 298, 302, 303, 306, 307, 311, 325, 346, 347, 349
allo-isoleucine, 298, 347
isopentenylpyrphosphoric acid, 362
isoprene, 361
isopropyl ester, 115
isosaccharino-1,4-lactone, 162
isotacophoresis, 158
isothiocyanate, 88
isothiocyanate derivatization, 98
isotope peak, 18
isovaleric acid, 296, 297, 314, 315, 316, 317, 322
isovaleric acidemia, 5, 294, 297, 314, 315, 316
isovaleric aciduria, 84
isovaleryl-β-D-glucuronide, 315
N-isovalerylalanine, 297, 315, 316, 317
isovaleryl-β-D-glucuronide, 297, 316, 317
isovalerylcarnitine, 296, 297
isovaleryl-CoA, 194, 308, 315, 316, 327, 329, 346
isovaleryl-CoA dehydrogenase, 308, 314, 315, 322, 370, 373
N-isovalerylglutamic acid, 297, 315, 317
N-isovalerylglutamine, 297, 317
isovalerylglycine, 296, 314, 316, 324, 327, 367, 373
N-isovalerylglycine, 297, 315, 316
N-isovalerylsarcosine, 297, 315, 316, 317
ITMS, 63, 65, 68

Jamaican vomiting sickness, 365, 372, 373
jet separator, 5, 54
joint pain, 163

keto acid, 77, 82
keto bile acid, 271
keto groups, 110
3-keto-Δ^4-steroid 5α-reductase, 260
3-keto-Δ^4-steroid 5β-reductase deficiency, 256, 258
2-keto-3-methylvaleric acid, 193, 194, 295, 298, 303, 325, 346, 347, 349
4-keto-6-hydroxyheptanoic acid, 301, 339, 340
4-keto-6-hydroxypheptanoic acid, 337
ketoacidosis, 194, 302, 324, 355
2-ketoadipic acid dehydrogenase, 330, 332
2-ketoadipic acid, 282, 298, 332, 333
2-ketoadipic acidemia, 330
2-ketoadipic aciduria, 298, 330, 332
11-ketoandrosterone, 161, 181
2-ketobutyric acid, 193, 349, 355
4-ketobutyric acid, 193
ψ-ketocarboxylic acid, 367
2-ketocarboxylic acid, 194
keto-enol tautomerism, 306, 332
11-ketoetiocholanolone, 161, 181
2-ketoglutaramic acid, 284
α-ketoglutarate dehydrogenase complex, 348
2-ketoglutaric acid, 159, 199, 282, 295, 297, 298, 304, 332, 347, 348, 349, 351, 354, 355, 356
3-ketohexanoic acid, 193, 200
5-ketohexanoic acid, 193, 367
2-ketoisohexanoic acid, 193, 194, 295, 298, 315, 327, 329, 346, 347, 349
2-ketoisovaleric acid, 193, 194, 295, 298, 346, 347, 349
7-ketolithocholic acid, 256, 262
ketone, 87, 191, 192, 200, 280
ketone body, 199, 314
7-ketooctanoic acid, 367
6-keto-$PGF_{1\alpha}$, 100, 102, 103

5-ketoproline, 343
β-ketothiolase deficiency, 298, 324, 325
β-ketothiolase, 324, 325
ketotic dicarboxylic acid, 365
ketotic dicarboxylic aciduria, 367
ketotic hyperglycinemia, 324
2-ketovaleric acid, 193
3-ketovaleric acid, 199, 299, 300, 303, 307, 313, 361
3-ketovaleryl-CoA, 303, 307
kidney, 157
kidney transplantation, 163
kinetic energy, 65
Kussmaul breathing, 192

lactate dehydrogenase, 126, 357
lactic acid, 87, 162, 193, 196, 197, 295, 296, 298, 299, 304, 308, 313, 324, 327, 348, 349, 351, 361, 367, 373
lactic acidemia, 350
lactic acidosis, 302, 348, 349
β-lactoglobulin, 130
lactyllactate, 304
LADH, 217
laser desorption (LD), 4, 30
laser desorption ionization (LDI), 30
LC, 8
LC/MS, 7, 8, 24, 25, 56, 59, 61, 75, 83, 86, 94, 99, 106, 107, 114, 125, 134, 156, 157, 158, 177, 179, 180, 191, 228, 244, 274, 275
LC/MS/MS, 107, 110, 137
LCAD deficiency, 369, 373
LCHAD deficiency, 369
lethargy, 192, 302, 310, 314, 324, 328, 346, 360, 363, 368, 369
leucine, 120, 192, 194, 254, 280, 281, 295, 298, 308, 314, 315, 326, 327, 328, 346, 347, 349
leukopenia, 314
leukotriene, 100
leukotriene B_4, 104
linear, 36
linkage analysis, 94
linked scan, 66, 210
linked scan method, 65
lipid storage myopathy, 369
lipid-bound, 181
Lipidex 5000, 109
Lipidex 5000 microcolumn, 110
Lipidex-DEAP, 277
lipophilic Sephadex, 109
lipoxin, 104
liquid chromatography (LC), 6
liquid chromatography/mass spectrometry (LC/MS), 56
liquid SIMS (LSIMS), 4, 27
lithocholic acid, 255, 256, 260, 269, 272
liver, 253, 254
liver alcohol dehydrogenase (LADH), 216
liver cirrhosis, 264, 265, 266, 270, 277, 281, 282, 337
liver disease, 271
long-chain 3-hydroxyacyl-CoA dehydrogenase (LCHAD) deficiency, 368, 369
long-chain acyl-CoA dehydrogenase (LCAD) deficiency, 294, 368, 369
low energy CID, 66
LSIMS, 6, 8, 23, 24, 26, 27, 41, 46, 48, 64, 168, 169, 271, 293
LSIMS/MS, 8, 294
lysine, 120, 206, 213, 308, 318, 319, 332
lysyl endopeptidase, 119, 124

$M^{-\cdot}$, 16
$M^{+\cdot}$, 16
macrocephaly, 317
magnetic, 210
magnetic field, 31, 32, 54, 66
magnetic field strength, 39
magnetic scanning, 54
magnetic sector, 20, 31, 43, 65, 125
magnetic sector analyzer, 31
magnetic sector mass analyzer, 32
magnetic sector mass spectrometer, 3
magnetic sector MS, 31
Maillard, 206
Maillard reaction, 206, 209
makeup gas, 54
MALDI, 8, 30, 31, 119, 130
MALDI-MS, 119, 129, 139, 156
MALDI-TOF, 130
MALDI-TOFMS, 31, 129, 130, 140
maleic acid, 301

maleylacetoacetic acid, 337, 338, 342
malic acid, 159, 162, 304, 350
malignant disease, 131
mannitol, 161, 174, 175, 177, 178, 179, 180, 201, 205
MAO, 228, 230
MAO-B inhibitor, 226
maple syrup urine disease, 294, 298, 345, 346, 347
mass analyzed ion kinetic energy (MIKE), 65
mass analyzed ion kinetic energy spectrometry (MIKES), 7
mass analyzer, 15, 31
mass chromatogram, 56, 86, 177, 179
mass chromatography (MC), 55, 110
mass fragmentography, 5
mass selective instability scan, 37
mass spectrometry (MS), 3
mass spectrum, 16
mass-to-charge ratio (*m/z*), 15
matrix, 25, 26, 27, 30, 61, 139
matrix-assisted laser desorption (MALD), 30
matrix-assisted laser desorption ionization (MALDI), 4, 30
MCAD deficiency, 368, 369, 373
McLafferty-type rearrangement, 306
medium-, short-, and long-chain acyl-CoA dehydrogenase, 322
medium-chain acyl-CoA dehydrogenase (MCAD) deficiency, 368, 369
medium-chain dicarboxylic acid, 192, 368, 370
medium-chain triglyceride milk, 373
medium-chain triglycerides, 365
megaloblastic anemia, 310
melanin, 334
mental and physical retardation, 346
mental retardation, 310, 324, 334, 341, 343, 349
mesaconic acid, 297, 304, 315, 316
metabolic acidosis, 318, 322, 326, 328, 330, 343, 345, 346, 358, 368, 369
metabolic ketoacidosis, 310, 314, 360
metabolic profiling, 6, 7, 199
metanephrine, 96
metastable, 18
metastable ion, 17, 66
metastable peak, 18
methane, 20, 22, 40, 117
methionine, 120, 199, 254, 280, 302, 303, 311, 338, 355
methoximation, 77, 83, 104
methoxime, 77, 78, 83, 101, 110
3-methoxy-4-hydroxyphenylethanol (MHPE), 246
3-methoxy-4-hydroxyphenylethyleneglycol (MHPG), 246
3-methoxy-4-hydroxyphenylpyruvic acid, 245
3-methoxycatechol, 160, 170, 171, 173
methoxyhydroquinone, 160, 170, 171, 173
methoxylamine hydrochloride, 101
2-methoxyresorcinol, 160, 170, 171, 172, 173, 174
methyl, 92
methyl boric acid, 94
methyl ester, 115, 158
methyl esterification, 117
methyl ester-MO-TMS, 105
methyl ester-TFA derivative, 116
methyl sulfinyl carbanion, 93
methyl *tert*-butyl ether, 89
N-methyl-1,2,3,4-tetrahydroisoquinoline (NMTIQ), 230
2-methyl-1-butanol, 201
3-methyl-1-butanol, 201
2-methyl-1-propanol, 201
24-methyl-26,27-dinor-5β-cholestane-3α,7α,12α,24-tetrol, 278
5(3)-methyl-3(5)-isoxazole propionic acid, 341
2-methyl-3-hydroxybutyric acid, 298, 299, 300, 303, 304, 313, 324, 325, 326
2-methyl-3-hydroxybutyric aciduria, 298, 324
2-methyl-3-hydroxybutyric-CoA, 325
2-methyl-3-hydroxybutyryl-CoA, 303, 307, 324, 325
2-methyl-3-hydroxyvaleric acid, 300, 303, 304, 307
2-methyl-3-ketovaleric acid, 299, 300, 303, 307, 361

2-methyl-3-ketovaleryl-CoA, 303, 307
1-methyl-4-phenyl-1,2,3,6-tetrahydropyridine (MPTP), 223
1-methyl-4-phenyl-2,3-dihydropyridine (MPDP), 228
1-methyl-4-phenyl-2,3-dihydropyridinium ion ($MPDP^+$), 225
1-methyl-4-phenyl-4-propionoxy-piperidine (MPPP), 224
1-methyl-4-phenylpyridinium ion (MPP^+), 225
1-methyl-6,7-dihydroxy-1,2,3,4-tetrahydroisoquinoline, 243
2-methyl-6,7-dihydroxy-1,2,3,4-tetrahydroisoquinoline, 241
2-methyl-6,7-dihydroxy-isoquinolinium ion ($MDIQ^+$), 244
2-methyl-6-ethylpyrazine, 201
7-methyl-8-hydroxyguanine, 136
2-methylacetoacetic acid, 298, 299, 300, 303, 304, 306, 307, 313, 324, 325
2-methylacetoacetic aciduria, 324
2-methylacetoacetyl-CoA, 303, 306, 324, 325
methylacrylyl-CoA, 194
1-methyladenosine, 131
N^6-methyladenosine, 131
3-methyladipic acid, 159, 162, 282, 299, 301, 304, 308, 313
methylation, 80, 82, 133, 164
N-methyl-bistrifluoroacetamide (MBTFA), 98
3-methylbutanal, 281
3-methylbutrylglycine, 303
2-methylbutyric acid, 296, 322
methylbutyryl-CoA dehydrogenase, 322, 370
2-methylbutyryl-CoA, 194, 303, 308, 325, 346
2-methylbutyrylglycine, 296, 301, 308, 324, 367
methylcitric acid, 299, 300, 303, 304, 305, 306, 313, 314, 325, 327, 330, 361
methylcobalamin, 311
methylcobalamin synthesis deficiency, 311
3-methylcrotonyl-CoA, 194, 315, 326, 327, 329
3-methylcrotonyl-CoA carboxylase, 308, 324, 326, 327, 360
3-methylcrotonyl-CoA carboxylase deficiency, 298, 326, 327
3-methylcrotonylglycine, 297, 298, 299, 327, 328, 329, 361
3-methylcrotonylglycine-CoA, 327
3-methylcrotonylglycinuria, 298, 326
5-methylcytosine, 136, 359
N-methyldopamine, 96
3,4-methyleneadipic acid, 159, 282
methylenecyclopropylacetic acid, 373
3,4-methylenesuberic acid, 282
N-methylepinephrine, 95, 99
5-methylfurancarboxylic acid, 282
2-methylglutaconic acid, 298, 299, 301, 308, 313, 324, 325
3-methylglutaconic acid, 282, 297, 298, 301, 304, 308, 327, 329, 330, 331, 355, 356
3-methylglutaconic aciduria, 298, 355, 356
3-methylglutaconyl-CoA hydratase, 355, 356
3-methylglutaconyl-CoA, 315, 326, 327, 329, 356
2-methylglutaric acid, 159
3-methylglutaric acid, 159, 282, 297, 298, 329, 355, 356
2-methylglyceric acid, 159, 162
methylguanidine, 156
1-methylguanosine, 131
N^2-methylguanosine, 131
methylhydroxylamine hydrochloride, 78
O-methylhydroxylamine hydrochloride, 77, 83, 104, 105
1-methylinosine, 131
N-methylisoquinolinium ion ($NMIQ^+$), 235
D-methylmalinonyl-CoA, 303
L-methylmalinonyl-CoA, 303
methylmalonic acid, 86, 87, 282, 299, 310, 312, 313, 325
methylmalonic acidemia, 310
methylmalonic aciduria, 84, 86, 294, 299, 302, 308, 310, 311, 312, 314, 321, 325
methylmalonylcarnitine, 299
methylmalonyl-CoA, 302
methylmalonyl-CoA mutase, 302, 311

methylmalonyl-CoA racemase, 302, 311
D-methylmalonyl-CoA, 302, 311, 312
L-methylmalonyl-CoA, 302, 311, 312
methylmercaptan, 160
N-methyl-*N*-nitroso-*p*-toluenesulfonamide, 113
3′-*O*-methylnorlaudanosolinecarboxylic acid, 245
N-methylnorsalsolinol, 96, 99, 241, 242, 243, 244
N-methyl-*N*-*tert*-butyldimethylsilyltrifluoroacetoamide (MTBSTFA), 81
N-methyl-*N*-trimethylsilylacetamide (MTMSA), 80
N-methyl-*N*-trimethylsilyltrifluoroacetamide (MSTFA), 79, 81
methyloxime, 83
O-methyloxime-TMS derivative, 94
O-methyloxime-TMS derivatization, 94
4-methylpimelic acid, 299, 301, 308, 313
7-methylpurine, 135
methylpyrazine, 201
N-methyl-salsolinol, 96, 99, 241, 242, 243, 244
3-methylsuberic acid, 159
4-methylsuberic acid, 299, 301, 308, 313
methylsuccinic acid, 162, 295, 296, 297, 304, 315, 316, 322, 366, 370, 373
methylsulfinyl carbanion method, 135
2-methylthiofuran, 201
1-methyl-TIQ, 230, 239, 241
N-methyltransferase, 235, 243
methyltrifluoroacetamide (IPDMSMTFA), 99
3-methyluridine, 133, 135
mevalonate kinase, 361, 362
mevalonic acid, 299, 329, 361, 362, 363
mevalonic aciduria, 299, 361, 362
mevalonolactone, 299, 361, 362
MHPE, 246
microcephaly, 352, 358
microcolumn, 60
β_2-microglobulin (β_2-MG), 157
middle molecule, 180
middle molecule fraction, 157
mitochondria, 373
mitochondrial β-oxidation, 368
mitochondrial cobalamin adenosyltransferase deficiency (cblB), 311
mitochondrial state 3 respiration, 169
m-nitrobenzylalcohol, 26, 27
MO, 83
modified headspace sampling, 90
modified nucleoside, 131
modified ribonucleoside, 133
molecular ion, 15, 16, 18, 19, 20, 22, 23
molecular SIMS, 27
monoamine oxidase (MAO), 225
mononucleotide, 134, 135
MO-pentafluorobenzyl (PFB), 104
MO-PFB, 104
MO-TMS, 83, 108, 109, 110, 207, 208, 304, 340
moving belt interface, 6
MPDP, 227
$MPDP^+$, 228
MPP^+, 227, 228, 230, 235
MPPP, 225
MPTP, 224, 225, 226, 227, 228, 229, 230, 231, 241, 243
MPTP lactam, 228, 229
MPTP pyridone, 228, 229
MPTP-*N*-oxide, 228
MS/MS, 8, 16, 38, 40, 64, 65, 67, 106, 119, 138
MS/MS (tandem mass spectrometry), 7
MS/MS/MS (MS^3), 66, 68
MS/MS/MS/MS, 68
MS^n, 66
MTBSTFA, 116, 136
multiple acyl-CoA dehydrogenase, 322
multiple carboxylase deficiency, 299, 359, 360, 361
multiple stages of MS/MS (MS^n), 68
multiply charged ion, 64
multiply charged molecular ion, 28, 30, 61, 125
multiply charged peptide ion, 119
ω-muricholic acid, 271, 272
muscle weakness, 369
muscular hypotonia, 355
mut^-, 311
mutase apoenzyme deficiency (mut°), 311

myelin, 371
myoclonal jerks, 358
myoglobin, 130
myoglobinuria, 370
myo-inositol, 156
myopathy, 174

N_2 laser, 129
Na,K-ATPase, 182, 183, 184
NADH dehydrogenase, 226
NaI, 50
natural abundance, 43
natural cyclotron frequency, 39
nausea, 192
negative ion APCI-LC/MS, 364
negative ion chemical ionization mass spectrometry (NICIMS), 40
negative ion mass spectrometry (NIMS), 40
neonatal adrenoleukodystrophy, 261, 262, 372
neonatal leukodystrophy, 365
neonatal tyrosinemia, 337
nephropathy, 209
nephrosclerosis, 155
neurological dysfunction, 156
neurological impairment, 358
neurotoxicity, 228
neurotoxin, 175, 225, 235
neutral fragment, 64
neutral gas, 66
neutral loss scan, 99
NICI, 105, 285
NICIMS, 41, 99, 104, 105, 117, 241, 270
NICI-SIM, 106
nicotinic acid, 30, 139, 140
NIMS, 41
NMR, 213
NMTIQ, 231, 235, 236, 237, 243
non-insulin-dependent diabetes mellitus (NIDDM), 191, 204, 205
non-ketotic dicarboxylic aciduria, 299, 365, 367, 368
non-ketotic hypoglycemia, 328, 369
non-paralytic esotropia, 351
nonylphenylpolyethylene sulfate, 49, 52
27-nor-5β-cholestane-3α,7α,12α, 25,26-pentol, 277, 278
27-nor-5β-cholestane-3α,7α,12α,24,25,26-hexol, 277, 278
27-nor-5β-cholestane-3α,7α,12α,24,25-pentol, 277
27-nor-5β-cholestane-3α,7α,12α,24,25-pentol, 278
nor-cholic acid, 272
norepinephrine, 96, 98, 223, 224, 245, 246
norgestrel, 108
norlaudanosolinecarboxylic acid, 245
normal serum, 162
normal-phase silica Sep-Pak cartridge, 100
normetanephrine, 96
nor-MPTP, 228
norsalsolinol, 241, 243, 244
nuclear magnetic resonance (NMR) spectroscopy, 7
nuclease P_α, 134
nucleic acid, 131, 134
nucleoside, 133, 134, 135, 136, 137, 138, 139
nucleoside triphosphate, 138
nucleotide, 133, 137, 139

obstructive biliary disease, 253
obstructive jaundice, 264, 265
ochronosis, 341, 342
octadecadienedioic acid, 366, 373
octadecanedioic acid, 366, 373
octadecenedioic acid, 366, 373
octadecylsilane (ODS), 100
octanoic acid, 369
1-octanol, 201
octanoylcarnitine, 295, 296, 300, 367, 368
octanoyl-CoA, 369
octanoylglucuronide, 300, 369
octanoylglycine, 300, 367, 368, 369
octenedioic acid, 301, 371, 373
2-octenedioic acid, 366
4-octenedioic acid, 366
octenoylcarnitine, 367, 368
octopamine metabolite, 246
ocular abnormality, 261
odd-chain fatty acid, 199
odd-numbered dicarboxylic acid, 372
odd-numbered fatty acid, 302, 303, 311
ODS column, 111, 131

ODS-bonded silica, 107, 111
oil diffusion pump, 15
oligonucleotide, 138, 139, 140
oligosaccharide, 92, 93
O-methylhydroxylamine hydrochloride, 110
one-stage jet separator, 50, 54
open-split coupling, 51, 53
optic atrophy, 355
optical nerve dysplasia, 371
organic acid, 6, 76, 157, 159, 161, 162, 191, 192, 193, 196, 197, 199, 254, 282, 293, 304, 305, 312, 316, 319, 322, 325, 329, 332, 335, 339, 341, 343, 345, 347, 349, 351, 354, 355, 357, 358, 359, 361, 364, 365, 372
organic acid metabolism, 7, 9
organic aciduria, 78, 86, 293, 294, 310, 314
organic solvent, 100, 107
ornithine, 363
ornithine transcarbamylase (OTC) deficiency, 84, 86, 300, 363
orotic acid, 86, 300, 363, 364
orotic aciduria, 300
osteoporosis, 351
OTC deficiency, 364
ouabain, 184
ouabain displacing digitalis-like factor, 183
ovalbumin, 126
oxalic acid, 159, 161, 163, 300, 356, 357
oxaloacetic acid, 305, 306
β-oxidation, 191, 192, 195, 199, 368, 369, 373
(ω-1)-oxidation, 199, 399
oximation, 77, 82, 83
oxime, 77, 82
oxime-TMS, 83
3-oxoacyl-CoA thiolase deficiency, 263
5-oxoprolinase, 344

packed column, 50, 54, 91
packed GC column, 87
palm and sole erosion, 341
parathyroid hormone, 180
parent ion, 63
Parkinson's disease, 6, 9, 223, 224, 225, 230, 232, 238, 241, 242, 244
parkinsonian, 241
parkinsonism, 224, 225, 233, 241
particle beam, 58
particle beam (PB) LC/MS, 56
PB, 110
PB-LC/MS, 58, 111
p-cresol, 156
PD, 24, 25
PDMS, 25, 228
PEG, 47, 48, 49, 50, 51, 52
pentadecanedioic acid, 366, 372
pentafluorobenzoyl chloride (PFBCl), 99
pentafluorobenzyl bromide (PFBB), 104
pentafluoropropionyl (PFP), 96
pentafluoropropionyl anhydride (PFPA), 96
pentafluoropropionyl-1,2,4-triazole (PFPT), 98
pentafluoropropionylimidazole (PFPI), 97
pentahydroxycholestanoic acid, 263
1-pentanol, 201
2-pentanone, 200, 201
3-pentanone, 301, 303, 307
3-pentene-2-one, 200
pentose, 213
pentosidine, 213, 214
peptidase, 344
peptide, 119, 125, 139
peptide CID mass spectrum, 121
perfluoro-2-propoxypropionylchloride (PPP-Cl), 239
perfluoroacylation, 98
perfluoroalkyl phosphazine, 46, 47
perfluorokerosene (PFK), 44
perheptafluorobutyrylimidazole, 98
peripheral neuropathy, 174
permethylation, 93
O,N-permethylation, 135
peroxisomal bifunctional protein, 263
peroxisomal β-oxidation, 372
peroxisomal β-oxidation enzyme, 262
peroxisomal disorder, 261, 262, 371, 372
peroxisomal enzyme alanine:glyoxylate aminotransferase, 163
peroxisomal enzyme D-glycerate dehydrogenase, 163
peroxisome, 261
PFB, 266, 270
PFB derivatization, 104

PFBB, 102, 103
PFK, 45, 46
PFP, 98, 115, 245
PFPA, 97, 116
PG, 99, 100, 102, 103, 105, 106, 107
PGD_2, 102
PGE_2, 102, 107
$PGF_{2\alpha}$, 102, 107
phenol, 156, 160, 170, 172, 173, 174, 224
phenolic acid, 161
phenolic carboxylic acid, 367
phenolic compound, 281
4-phenyl-1,2,3,6-tetrahydropyridine, 228
N-phenylacetyl-α-aminoglutarimide, 158
phenylacetic acid, 282, 300, 334, 335
phenylacetylglutamic acid, 158, 160
phenylacetylglutamine, 158, 160, 300, 334, 335
phenylalanine, 120, 254, 280, 300, 334, 336, 338, 341, 342
phenylalanine hydroxylase, 334
phenylalanine/tyrosine, 336
phenylboric acid, 94
phenylboronate affinity column, 131
phenylboronic acid cartridge (PBA), 102
phenylboronic acid column, 96, 101
phenylethanolamine *N*-methyltransferase, 223
β-phenylethylamine, 98
2-phenylethylamine, 231
phenylethylene glycol, 300, 334, 335
phenylketonuria, 84, 85, 86, 117, 294, 300, 334, 335, 336
phenyllactic acid, 85, 86, 300, 301, 334, 335
phenylpropionylglycine, 367, 368
4-phenylpyridine, 228
phenylpyruvic acid, 85, 86, 300, 301, 334, 335, 336
phosphate, 363
phosphatidylinositol, 180
phosphatidylinositol diphosphate, 180
phosphatidylinositol monophosphate, 180
phosphodiesterase, 134
phosphoinositol, 180, 181
5-phosphomevalonic acid, 361, 362
phosphorylated amino acid, 121
phosphorylation, 130
PHP-LH-20, 111
PHP-LH-20 column, 112
phytanic acid, 199, 302, 311, 371, 372
picoline, 201
Pictet-Spengler condensation, 243, 244, 245
Pictet-Spengler reaction, 231
pigmentary retinopathy, 371
pimelic acid, 159, 162, 282, 299, 301, 308, 313, 366, 372
pipecolic acid, 371, 372
piperidinohydroxypropyl Sephadex LH-20 (PHP-LH-20), 277
piperidinohydroxypropyl-LH-20 (PHP-LH-20), 109
plaque, 341
plasma desorption (PD), 4, 24
plasma desorption mass spectrometry (PDMS), 24
plasmalogen, 371, 372
plasmaspray LC/MS, 314
platelet function, 156
pneumatically assisted ESI, 29, 62
polyamine, 156
polycystic kidney disease, 155
polyethyleneglycol (PEG), 46, 47
polyhydric alcohol, 174
polymorphism, 373
polyol, 91, 92, 93, 94, 161, 174, 175, 176, 177, 179, 191, 192, 201, 202, 208
polysaccharide, 94
Porasil E, 90
porphyria, 338
porphyrin, 354
post source decay (PSD), 63
potassium adduct ion, 23, 25
PPP, 241
prealbumin, 120
prealbumin variant, 8, 123
precursor ion, 55, 63, 64, 68, 210, 213
precursor ion scan, 65, 67
precursor ion spectrum, 67
primary biliary cirrhosis, 271, 272, 277
primary hyperoxaluria, 163, 356
primary hyperoxaluria type 1, 300, 357
primary hyperoxaluria type 2, 300, 357
primary hyperoxaluria type I, 163
primary hyperoxaluria type II, 163

product (daughter) ion spectrum, 121
product ion, 7, 55, 63, 64, 65, 66, 67, 68, 122, 213
product ion scan, 65, 67
product ion spectrum, 64, 65, 67
profound hypoglycemia, 372
proline, 295, 348, 349
1-propanol, 201
2-propanol, 201
propionic acid, 199, 299, 300, 302, 303, 305, 306
propionic acidemia, 294, 300, 302, 303, 308, 313, 321, 325, 330
propionic aciduria, 84
propionylcarnitine, 299, 301, 309, 310
propionyl-CoA, 199, 302, 303, 305, 307, 308, 309, 311, 312, 313, 314, 324, 325, 355
propionyl-CoA carboxylase, 302, 303, 311, 360
propionylglycine, 300, 303, 308, 325, 330
n-propyl ester, 115
prostaglandin (PG), 5, 99
protein, 119, 124, 125
protein binding of drugs, 163, 164
protein carboxyl methyltransferase, 125
protein glycosylation, 129
protein-bound acid, 167
protein-bound metabolite, 164, 169
protein-bound organic acid, 163
protein-losing gastroenteropathy, 354
proton abstraction, 41
proton transfer, 20, 22
protonated free base ion, 137
protonated molecular ion, 22, 23, 24, 25, 27, 30, 42, 57, 59, 64, 137, 228
protoporphilin IX, 253
proximal tubule, 157
pseudouridine, 131, 133, 137, 138, 158
psychomotor retardation, 330, 351, 352, 355, 356, 358, 361
pulsed laser ablation, 140
pyrazine, 201
pyridine, 160, 201
pyrimidine pathway, 363
pyroglutamase, 344
pyroglutamic acid, 284, 301, 343, 344, 345
pyroglutamic aciduria, 301, 343, 344, 345
2-pyrolidone-5-carboxylic acid, 158
5-pyrphosphomevalonic acid, 362
pyrraline, 211, 213
pyrrole, 87
2-pyrrolidone-5-carboxylic acid, 284
pyruvate carboxylase, 359
pyruvate carboxylase deficiency, 349
pyruvate dehydrogenase, 353
pyruvate dehydrogenase complex, 348
pyruvic acid, 199, 282, 295, 298, 348, 349, 351, 373
pyruvic acid decarboxylase, 302
pyruvic acidemia, 350

QMS, 37, 38, 63
quadrupole, 4, 43, 65, 226
quadrupole mass filter, 4, 33, 35
quadrupole mass spectrometer, 125
quadrupole mass spectrometry (QMS), 33
quinoiddihydrobiopterin, 224

radio frequency (RF) voltage, 33
reactant gas, 20, 22, 40, 57
reagent ion, 20, 21
recombinant human methionyl growth hormone, 125
reconstructed ion chromatogram (RIC), 56
reduction, 94
reflector, 36
relative abundance, 16
relative intensity (abundance), 15
renal cyst, 371
replacement by $O^{-\cdot}$, 40, 41
resolution, 42, 44
resonant ejection, 38
resorcinol, 160, 171, 172, 173
retardation of development, 326
reverse geometry, 7, 32
reverse geometry double-focusing mass spectrometer, 67
reverse-phase C_4 column, 125
reverse-phase cartridge, 100
Reye's syndrome, 294, 365, 368, 373
Reye-like illness, 322
Reye-like syndrome, 365, 373
RF voltage, 34, 37, 38, 66

ribitol, 175, 178
riboflavin, 318, 322
ribonuclease, 216, 217
ribonucleoside, 131, 133, 138, 139
ribose, 213
1-ribosylpyridine-4-one-3-carboxamide, 131
Richner-Hanhart syndrome, 341
rickets, 338
ring electrode, 37
RNA, 133, 134, 137
RNase A, 134
RNase T_2, 134

salsolinol, 241, 243, 245
sarcosine, 316, 324
sarcosine dehydrogenase, 322, 370
saturated dicarboxylic acid, 366
SCAD deficiency, 369
SCHAD deficiency, 370
Schiff base, 206
Schwann cell, 181
sclerosing cholangitis, 271
SCX, 177
sebacic acid, 192, 193, 282, 295, 296, 299, 322, 351, 366, 367, 368, 370, 372, 373
sebacylcarnitine, 296, 367
second generation product ion, 68
secondary ion mass spectrometry (SIMS), 4, 27
sector type, 65
sector type MS/MS, 66
seizure, 261, 302, 310, 311, 314, 317, 352, 358, 360, 372, 373
selected ion monitoring (SIM), 5, 38, 54
selected reaction monitoring (SRM), 55
self-CI, 40
semimicrocolumn, 60
separator, 54
Sephadex DEAE A-25 column, 78
Sephadex G-10, 95
Sephadex LH-20 column, 106, 110
Sep-Pak C_{18}, 100, 107, 108, 109, 111, 277
Sep-Pak C_{18} cartridge, 112, 113, 277
serine, 358
serum alkaline phosphatase (ALP), 253
sheath gas, 29
short-chain 3-hydroxyacyl-CoA dehydrogenase (SCHAD) deficiency, 368, 370
short-chain acyl-CoA dehydrogenase (SCAD) deficiency, 368, 369
short-chain fatty acid, 86, 87
short-chain hydrogen carbon, 88
shortening of the life span of red blood cells, 161
silic acid column chromatography, 104
silic acid partition chromatography, 86, 87
SIM, 55, 56, 99, 105, 106, 110, 113, 114, 170, 173, 177, 199, 202, 204, 207, 237, 238, 239, 241, 242, 245, 246, 264, 265, 270, 277, 313, 314, 324, 341, 357, 362
SIM chromatogram, 55
SIMS, 27
sinapinic acid, 30, 31, 129, 130
single-focusing (direction-focusing), 3
single-focusing magnetic sector mass analyzer, 18
single-focusing mass spectrometer, 32
skin rash, 360
small protein, 157
snake venom exonuclease, 134
sodium adduct ion, 23, 25
sodium boron hydride, 94
sodium iodide, 48
sodium octyl sulfate, 119
soft ionization, 4, 8, 23, 41, 61, 64, 129
solid-phase extraction, 100
solvent extraction, 76, 87, 88, 89, 90
solvolysis, 112
sorbitol, 161, 174, 175, 177, 178, 180, 205
spastic paraparesis, 355
spastic quadriplegia, 343
spastic tetraparesis, 358
spasticity, 351
speech delay, 358
speech retardation, 356
spermine, 156
spleen exonuclease, 134
splitless injection method, 161
SRM, 64, 106, 110, 114
stable isotope, 5, 18, 55
stable isotope dilution GC/MS, 362, 364
stable isotope dilution method, 55, 119, 313, 339, 341

stable isotope dilution MS, 357
stable isotope labeled amino acid, 115
stable isotope labeled compound, 106, 110, 114
stable isotope labeled internal standard, 98, 138
Staphylococcus V_8 protease, 119
static SIMS, 27, 28
steam distillation, 86, 87
steroid, 6, 107, 161, 181
steroid disulfate, 109
steroid glucuronide, 109
steroid monosulfate, 109
suberic acid, 192, 193, 282, 295, 296, 299, 301, 304, 322, 351, 353, 366, 367, 368, 370, 371, 372, 373
suberylcarnitine, 296, 367
suberylglycine, 296, 300, 324, 367, 368
succinic acid, 30, 140, 162, 193, 282, 295, 297, 304, 350, 351, 352, 355
succinic semialdehyde, 297, 352, 353, 354
succinic semialdehyde dehydrogenase, 352, 353
succinic semialdehyde dehydrogenase deficiency, 297
succinylacetoacetic acid, 301, 337, 339, 340
succinylacetone, 301, 337, 338, 339, 340
succinyl-CoA, 199, 302, 303, 311, 348
sudden death, 318
sudden infant death syndrome, 368
sugar, 91, 92, 93, 94
sugar ion, 137
sulfate, 99, 107, 109, 170, 174, 181
sulfate conjugate, 112
sulfate-conjugated bile acid, 111
sulfated amino acid, 121
sulfated bile acid, 265
sulfide, 88
sweaty feet odor, 314, 322
systemic carnitine deficiency, 301, 365, 370, 371

tandem mass spectrometry (MS/MS), 63, 210
tandem-in-space, 63, 65
tandem-in-time, 63, 65
tartaric acid, 158, 159
taurine, 255, 256, 258, 259, 262, 263
taurine conjugate, 112
taurine-conjugated bile acid, 111
taurochenodeoxycholate, 255
taurochenodeoxycholic acid, 275, 279
taurocholate, 255
taurocholic acid, 275, 279
taurodihydroxy, 273
taurodihydroxycholanoate, 114
taurohydroxy, 273
taurotrihydroxycholanoate, 114
Taylor cone, 29
TCA cycle, 303, 306, 311, 350, 352, 354
TEAP-LH-20, 111
Tenax GC, 88, 89, 91
tetradecadienedioic acid, 366, 373
tetradecanedioic acid, 366, 373
tetradecenedioic acid, 373
5-tetradecenedioic acid, 366
tetrahydrobiopterin, 224
tetrahydrocholanoic acid, 263
tetrahydrocortisone glucuronide, 184
tetrahydroisoquinoline (TIQ), 230
1,2,3,4-tetrahydroisoquinoline (TIQ), 97, 224, 230
tetrahydropapaveroline, 245
1β,3α,7α,12α-tetrahydroxy-5β-cholan-24-oic acid, 272
1β,3α,7α,12α-tetrahydroxy-5β-cholanoic acid, 266
24ξ,25ξ-3α,7α,12α,24-tetrahydroxy-5β-cholestan-26-oic acid, 261
(22*R*)-3α,7α,12α,22-tetrahydroxy-5β-cholestanoic acid, 261
(23*R*)-3α,7αtetrahydroxy-5β-cholestanoic acid, 12α,22-, 261
tetrahydroxycholanoate, 273
tetrahydroxycholestanoic acid, 263
TFA, 97, 98, 110, 113, 115, 270
TFA derivative, 93, 136
TFA derivatization, 93, 114
TFAA, 97, 110, 113, 116
THCA, 263, 371
thermospray (TSP), 25
thermospray ionization, 25
thermospray ionization (TSI), 6

thermospray ionization (TSI) LC/MS, 57
thiamine, 346
thiamine pyrophosphate, 346
thin-layer chromatography/mass spectrometry (TLC/MS), 56
thioglycerol, 26, 27
thiol group, 98
thionyl chloride, 116
threitol, 161, 175, 177, 178
threo-4,5-dihydroxyhexanoic acid, 354
threonic acid, 162
threonine, 302, 303, 311, 355
L-threonine, 199
N^6-threoninocarbonyladenosine, 131
thromboxanes (TXs), 99
thymidine, 136, 138
thymine, 295, 358, 359
TIC, 61, 177, 179
tiglic acid, 300, 303
tiglyglycine, 303
tiglylcarnitine, 298, 325
tiglyl-CoA, 194, 303, 308, 324, 325
tiglylglycine, 298, 299, 301, 308, 313, 324, 325, 361
timed ion selection, 63
time-of-flight (TOF), 3, 20, 25, 35
time-of-flight mass spectrometry (TOFMS), 36
TIQ, 231, 232, 233, 234, 235, 236, 237, 238, 239, 240, 241, 243, 245, 246
TLC, 104
TLC/FAB-MS, 56
TLC/LSIMS, 56
TLC/MS, 56
TMCS, 81, 99, 109, 113, 135
TMS, 78, 80, 91, 92, 99, 104, 105, 106, 109, 113, 170, 172, 173, 176, 181, 195, 198, 242, 243, 272
TMS derivatization, 110, 116, 135
TMSI, 81, 109, 110
TOF, 30, 37, 43, 125, 129
TOFMS, 36, 63, 140
toluene, 160
N-tosylphenylalanine chloromethyl ketone, 120
total ion chromatogram (TIC), 56
toxin, 254, 284
TPCK-trypsin, 120
2-*trans*, 4-*cis*-decadienoylcarnitine, 371
transaminase, 368
transevaporator sampling, 88, 90
transferrin, 128, 129
transmitochondrial malate shuttle, 302
tri(2,4-xylenyl)phosphate (TXP) column, 173
tricarboxylic acid (TCA) cycle, 199, 302
tridecanedioic acid, 366, 372
triethanolamine, 26, 27
triethylaminohydroxypropyl-LH-20 (TEAP-LH-20), 109
triethylammonium bicarbonate, 134
triethylammonium sulfate, 108
trifluoroacetic anhydride (TFAA), 93
trifluoroacetyl (TFA), 92
trifluoroacetylation, 93
trifluoroacetylimidazole (TFAI), 97
triglyceride, 351
3,16,17-trihydroxy-11-ketoandrostane, 161, 182
3α,7α,12α-trihydroxy-26,27-dinor-5β-24-one, 278
3α,7α,12α-trihydroxy-27-carboxymethyl-5β-cholestan-26-oic acid, 261
3β,7α,12α-trihydroxy-5α-cholanoic acid, 259
1β,3α,12α-trihydroxy-5β-cholan-24-oic acid, 272
1β,3α,7β-trihydroxy-5β-cholan-24-oic acid, 272
1β,3α,12α-trihydroxy-5β-cholanoic acid, 266
1β,3α,7α-trihydroxy-5β-cholanoic acid, 266
3α,6α,7α-trihydroxy-5β-cholanoic acid, 266
3α,7α,12α-trihydroxy-5β-cholestanoic acid, 261, 371
3β,7α,12α-trihydroxy-5-cholenoic acid, 256, 258
3,16,17-trihydroxyandrostane, 161, 182
trihydroxybile acid, 259
trihydroxycholanoate, 273
trihydroxycoprostanic acid (THCA), 261, 262
trimethylamine, 156
trimethylanilinium hydroxide (TMAH), 107

trimethylanilinium hydroxide method, 135, 136
trimethylchlorosilane (TMCS), 77, 80
2,3,5-trimethylpyrazine, 201
trimethylsilyl (TMS), 77
N-trimethylsilylacetamide (TMSA), 80
trimethylsilylated, 96
trimethylsilylation, 79, 81, 83, 105, 215
N-trimethylsilyldiethylamine (TMSDEA), 81
N-trimethylsilyldimethylamine (TMSDMA), 81
N-trimethylsilylimidazole (TMSI), 80
triple quadrupole, 35, 65, 66
triple quadrupole mass spectrometer, 121
tRNA, 131, 133, 134, 135, 137, 139, 140, 158
trypsin, 119, 124
trypsin digestion of protein, 129
trypsinogen, 130
tryptophan, 120, 158, 254, 280, 308, 318, 319, 332, 336
trytophan, 318
TSI, 25, 57, 110, 136
TSI-LC/MS, 25, 56, 57, 84, 99, 106, 107, 111, 114, 137, 275, 277, 279
turbomolecular, 15
TX, 100, 102, 104, 105
TXB_2, 101, 103, 106, 107
m-tyramine, 246
p-tyramine, 246, 337
tyrosine, 120, 224, 281, 282, 301, 334, 335, 336, 337, 338, 339, 341, 342
tyrosine hydroxylase, 223, 224, 230, 233, 235
tyrosine transaminase, 282, 337, 341
tyrosinemia, 294
tyrosinemia type 1, 301, 336, 337, 338, 339
tyrosinemia type 2, 301, 337, 341
tyrosinosis, 336

ubiquinone, 362
UDPG-glucuronyl transferase, 316
ultrafiltration, 79
ultraviolet (UV) spectroscopy, 7
undecanedioic acid, 366, 372
unsaturated 3-hydroxydicarboxylic acid, 366
unsaturated dicarboxylic acid, 366
uracil, 295, 300, 358, 359, 363, 364
urea, 162
urea cycle, 363
urease, 78, 79
β-ureidoisobutyric acid, 359
β-ureidopropionic acid, 359
uremia, 6, 9, 155, 157, 175, 181
uremic blood, 158
uremic encephalopathy, 174
uremic hemofiltrate, 182
uremic hemofiltrate and blood, 181
uremic patient, 133, 156, 161, 163, 177, 181
uremic peripheral neuropathy, 173, 180
uremic plasma, 182
uremic serum, 169, 177, 180
uremic serum ultrafiltrate, 158, 161, 162, 165
uremic symptom, 155, 161
uremic toxin, 155, 156
uric acid, 133, 158
uridine, 133, 137, 138
urobilinogen, 254
ursocholic acid, 272
ursodeoxycholic acid, 256, 260, 262, 269, 271, 272, 273, 275, 277

V_8 protease, 120, 124
vacuum distillation, 86, 87
valine, 192, 194, 254, 280, 295, 298, 302, 303, 311, 346, 347, 349
valley definition, 42
valley definition of 10%, 184
valproic acid, 365, 373
vanillic acid, 159, 282
vanilmandelic acid, 160
vaporizer probe, 57
vaporizer temperature, 179
varanic acid, 261, 262, 263, 371
velocity-focusing mass spectrometer, 3
venom phosphodiesterase, 134
very long chain fatty acid, 261, 262, 371, 372
VG 70SQ mass spectrometer, 183
vinylpyrazine, 201

viral hepatitis, 264
vitamin B_{12}, 311
vitamin B_2, 318
volatile, 87, 88, 89, 91, 160, 192, 200, 280
volatile compound, 254
vomiting, 192, 302, 310, 314, 317, 322, 324, 326, 343, 346, 348, 355, 360, 368, 369, 372, 373

white skin, 334
XAD-2, 107
XAD-7, 108
xylitol, 175
xylulose, 175

yellow-brown hair, 334

Zellweger (cerebrohepatorenal) syndrome, 261, 262, 365, 371, 372